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Racial incidence of coarctation of aorta[1]

R. L. van der Horst and M. S. Gotsman
From the Cardio-Thoracic Unit, Wentworth Hospital; and the Department of Medicine, University of Natal, South Africa

Published reports suggest that coarctation of the aorta occurs more frequently in certain racial groups. These impressions are probably fallacious, and are a consequence of the interpretation of data in which the patient samples exclude infants and undiagnosed cases and are therefore not representative of the total population sample. We analysed two separate groups of patients comparing four different races, and have shown that in clinically diagnosed cases, coarctation of the aorta appears to be more common in the White race. A necropsy diagnosis was made in 12 Bantu patients, suggesting that many more patients in this racial group remain undetected and that the true incidence of the lesion has been underestimated.

Coarctation of the aorta is a common congenital malformation and occurs in about 5 per cent of all patients with congenital heart disease. The mortality in relation to this lesion in infancy is high and is usually the consequence of multiple associated congenital cardiac malformations. If the analysis is confined to patients under the age of 1 year, the incidence is more than 10 per cent and in necropsy studies it may range from 10 to 21 per cent (Table 1).

Recent reports from Asia suggest that coarctation of the aorta is rare in certain racial groups. An analysis of these studies is summarized in Table 2. Similar reports from Africa suggest that the malformation is rare in African patients as well (Table 3).

We have not observed a racial difference in the spectrum of congenital heart disease (van der Horst *et al.*, 1968; van der Horst and Wainwright, 1969; van der Horst, Winship, and Gotsman, 1970) and similar observations have been made by Muir (1960), Phillips and Burch (1960), Watler (1960), Bradlow, Zion, and Fleishman (1964), Schrire (1964), Caddell and Connor (1966), Caddell and Morton (1967), and Mannheimer and Caddell (1968).

This paper will report our experience of the incidence of coarctation of the aorta in four racial groups living in Natal, South Africa. We feel that the reported racial difference in incidence is apparent and not real, and appears to be a result of circumstantial case selection and a consequence of failure to recognize this anomaly in infancy.

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Patients, methods, and results

An analysis was made of two separate groups of patients selected from two very different sources to show that coarctation of the aorta is not rare in the Bantu and to test the hypothesis that the racial difference in incidence is apparent and not real.

The first group of patients was derived from a series of subjects with heart disease who were admitted to the regional cardiothoracic centre and in whom a definitive diagnosis was established by cardiac catheterization with angiocardiography and/or operation.

FIG. *The ages and racial groups of the 33 patients with coarctation of the aorta (Cardiac Unit, Wentworth Hospital, 1963–1970).*

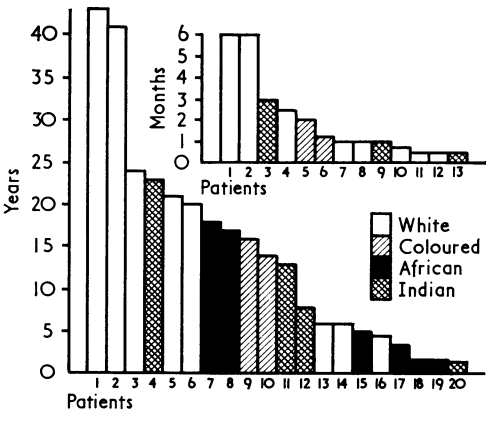

TABLE 1 *Series of congenital heart malformations reported from 'developed' centres with details of patients with coarctation of aorta*

Reference	*Abbott*	*MacMahon, McKeown, and Record*		*Kjellberg* et al.	*Carlgren*		*Nadas*	*Schrire*
Year	1936	1953		1959	1959		1963	1963
Type of study	Necropsy	Clinical, cath.	Necropsy and opn	Clinical, cath.	Clinical, cath.	Necropsy only	Clinical, cath., opn, necropsy	Clinical, cath., opn, necropsy
Age range	All ages	Newborn and infancy*	Newborn and infancy*	Mostly children	Newborn and infancy*	Newborn and infancy*	Mostly infants and children	0–69 yr
Cases of congenital heart disease	1000	117	225	742	224	145	3786	1439
Cases of coarctation of aorta	85	5	27	102	5	31	189	88
Per cent	8·5	4·3	12·0	13·7	2·2	21·3	4·9	6·0

* Serial studies on infants from birth.
Cath., cardiac catheterization with angiocardiography. NA, not admitted.

TABLE 2 *Series of congenital heart disease reported from Asian countries with details of patients with coarctation of aorta*

Reference	*Muir*	*Furuta*	*Inada* et al.	*Wada*	*Imperial and Felarca*	*Vakil*	*Sakakibara*	*Ongley*	*Vinijchaikul*
Year	1960	1961	1962	1963	1963	1963	1964	1966	1967
Country	Singapore	Japan	Japan	Japan	Philippines	India	Japan	Thailand	Thailand
Type of study	Necropsy	Opn, necropsy	NA	Opn	Necropsy	Clinical	Opn	Necropsy, opn	NA
Age range	All ages	NA*	NA	NA*	NA	Mostly older children and adults	<2 yr	NA	NA
Per cent infants <1 yr	NA	NA	NA	NA	NA	Few	NA	NA	NA
Cases of congenital heart disease	411	247	NA	332	67	500	90	NA	NA
Cases of coarctation of aorta	19	4	Very rare	1†	NA	16	6	1	Very rare
Per cent cases with coarctation	4·6	1·5	?	0·3	?	3·2	6·6	?	?

NA = Not admitted.
* Presumed older patients only
† Quoted by Shann but not detailed in paper in which 35 cases classified as 'other'.

TABLE 3 *Series of congenital heart disease reported from African countries with details of patients with coarctation of aorta*

Reference	*Caddell and Connor*	*Gupta and Antia*	*Caddell and Morton*	*van der Horst* et al.	*Wood, Serumaga, and Lewis*	*van der Horst and Wainwright*	*van der Horst* et al.
Year	1966	1967	1967	1968	1969	1969	1970
Country	Uganda	Nigeria	Nigeria	South Africa (Bantu)	Uganda	South Africa (Bantu)	South Africa (Indian)
Type of study	Clinical	Clinical, necropsy	Clinical	Cath., opn	Necropsy	Necropsy	Cath., clinical, opn
Age range	0–12 yr	0–1 yr	0–12 yr	All ages	All ages	All ages	All ages
Per cent infants <1 yr	60%	NA	NA	13%	45%	87%	21%
Cases of congenital heart disease	44	15	67	117	60	123	395
Cases of coarctation of aorta	0	1	0	1	5	3	9
Per cent cases of coarctation	—	7·3	—	0·8	8·3	2·4	2·3

Abbreviations: refer to Table 2.

Mehrizi, Hirsch, and Taussig	*Lambert, Canent, and Hohn*	*Gasul, Arcilla, and Lev*	*Keith, Rowe, and Vlad*	*Vince*	*Wood*	*Varghese* et al.	*Coleman*		*Krovetz, Gessner, and Schiebler*
1964	1966	1966	1967	1968	1968	1969	1969	1969 and 1965	1969
Necropsy	Necropsy	Clinical, cath., opn, necropsy	Clinical, cath., opn, necropsy	Cath.	NA	Cath.	Clinical, cath., opn, necropsy	Necropsy only	Clinical, cath., opn, necropsy
Newborn	Newborn	Mostly infants and children	Mostly infants and children	< 1 yr	Mostly beyond infancy	Newborn	< 1 yr	< 1 yr	0–15 yr
170	165	1943	6647	215	2000	100	380	177	2316
25	16	123	369	11	200	9	30	29	74
15·0	10	6·3	5·6	5·1	10	9·0	7·9	16·4	6·0

Shann	*Loh*		*Wallooppillai and Jayasinghe*
1969	1969		1970
Taiwan	Singapore		Ceylon
Cath.	Clinical, cath., opn, necropsy	Necropsy only	Cath., opn
All ages	< 20 yr	< 20 yr	All ages
12·5%	28% †	NA	4·9%
232	1184	289	555
3	17	12	4
1·3	1·4	4·2	0·7

† Estimated from Fig. 2 in Loh (1969).

The cardiothoracic unit is the regional centre for cardiac investigation and operation for all racial groups, and admits patients who will require operation or who present difficult diagnostic or therapeutic problems. The unit drains the Durban conurbation, whose municipal area in 1968 had a population of 187,778 White, 31,520 Coloured, 270,259 Indian, and 206,687 Bantu people. It also serves as the main hospital of referral for patients in the Province of Natal with a population of 337,409 White, 42,240 Coloured, 394,807 Indian, and 2,199,578 Bantu people. The number of patients submitted to cardiac catheterization, cases of proven congenital heart disease, and patients with coarctation of the aorta seen in the unit from 1 January 1963 to 1 September 1970 has been analysed and the results are given in Table 4. The racial distribution and the ages of the 33 patients with coarctation are shown in the Fig. Of the 33 patients, 16 were White, 4 were Coloured, 7 were Indian, and 6 were Bantu: patients with aortic arteritis are excluded. Details are shown in Table 5. From these data, it appears that coarctation of the aorta is most common in White subjects, and an analysis of the age spectrum of the patients shows that no African patient with this lesion was studied before the age of 18 months.

The second group of patients was obtained from an analysis of the necropsy records at King Edward VIII Hospital which is the main teaching hospital of 2099 beds for the

TABLE 4 *Analysis of cardiac catheterization procedures and racial incidence of congenital heart disease and coarctation of aorta from cardiac unit, Wentworth Hospital (1.1.1963 to 31.8.1970)*

Year	*Total No. of patients catheterized**	*No. of patients with congenital heart disease†*				
		Total	*White*	*Coloured*	*Indian*	*Bantu*
1963	111	66	14	2	23	27
1964	105	63	22	7	17	17
1965	109	54	20	2	16	16
1966	138	66	20	3	20	23
1967	170	106	34	7	29	36
1968	459	169	44	13	59	53
1969	523	171	59	17	53	52
1970 (8 months)	374	137	34	8	45	50
Total	1989	832	247	49	262	274
Coarctation of aorta			16	4	7	6

*Figures include repeat studies on a few patients performed for various reasons, i.e. follow-up, postoperative studies, additional data.
† Absolute figures.

TABLE 5 *Details of patients with coarctation of the aorta studied in the cardiac unit, Wentworth Hospital 1 January 1963 to 31 August 1970*

Case No.	Age	Sex	Other defects
White			
1	24 yr	M	Isolated
2	1 mth	M	Isolated
3	6 yr	M	Isolated
4	6 yr	M	Isolated
5	20 yr	F	Isolated
6	21 yr	M	Isolated
7	3 wk	F	Isolated
8	43 yr	M	Isolated
9	6 mth	F	Isolated
10	1 mth	M	Ventricular septal defect
11	41 yr	M	Mitral incompetence
12	4·5 yr	F	Atrial septal defect
13	2 wk	M	Atrial septal defect
14	2 wk	F	Atrial septal defect + pulmonary stenosis + persistent ductus arteriosus
15	10 wk	M	Atrial septal defect + ventricular septal defect + persistent ductus arteriosus
16	6 mth	M	Absent inferior vena cava + persistent ductus arteriosus
Coloured			
17	2 mth	F	Isolated
18	16 yr	M	Isolated
19	14 yr	M	Aortic stenosis
20	5 wk	M	Complete transposition of great vessels + ventricular septal defect + atrial septal defect + persistent ductus arteriosus
Indian			
21	23 yr	M	Isolated
22	3 mth	M	Persistent ductus arteriosus
23	1 mth	F	Persistent ductus arteriosus
24	8 yr	M	Persistent ductus arteriosus
25	13 yr	M	Aortic stenosis
26	2 wk	M	Atrial septal defect
27	16 mth	F	Dextrocardia; situs inversus; pulmonary stenosis; persistent ductus arteriosus
African			
28	17 yr	F	Isolated
29	3½ yr	M	Ventricular septal defect
30	19 mth	M	Persistent ductus arteriosus
31	19 mth	M	Persistent ductus arteriosus
32	18 yr	M	Isolated
33	5 yr	F	Isolated

Cases 21–26 in the Indian reported previously (van der Horst *et al.*, 1970). Case 28 in the African reported previously (van der Horst *et al.*, 1968).

Durban conurbation. The hospital admits patients of only the Indian and Bantu ethnic groups. White and Coloured patients are admitted to another Provincial Hospital, and a large proportion are treated in private nursing homes. During the period under review (1 January 1963 to 1 September 1970), a total of 11,970 necropsies was performed in patients of all age groups. A diagnosis of coarctation of the aorta was confirmed in 12 patients (those with significant hypoplasia of the left heart were excluded). Details of the age at death and the associated congenital abnormalities are given in Table 6. It is interesting to observe that a correct clinical antemortem diagnosis was made in only Case 12. The necropsy data give no indication of the true racial incidence of the lesion, as the hospital admitted patients of two racial groups only and also the relatives of many Indian patients were reluctant to permit necropsy examination. It does indicate that coarctation of the aorta occurs fairly frequently in the Bantu neonate, that they die of cardiac failure, and that the condition is clinically often unrecognized.

Discussion

Any analysis of published statistics which describes the incidence of coarctation of the aorta must recognize two important observations.

Firstly, coarctation of the aorta is relatively simple to diagnose in children and adults if the femoral and pedal pulses are examined as a routine in every patient. These observations are not new (Reifenstein, Levine, and Gross, 1947), but are not sought for routinely, particularly in children who have no symptoms referable to the cardiovascular system unless the attending doctor is aware of this physical sign, knows the relative frequency of the lesion, and has the time to make these

TABLE 6 *Coarctation of aorta in African: cases diagnosed at necropsy*

Case No.	1	2	3	4	5	6	7
Age at death	13 hr	24 hr	3 dy	4 dy	9 dy	14 dy	15 dy
Associated defects	Atrial septal defect, large	—	Persistent ductus arteriosus	Ventricular septal defect, single atrium, persistent ductus arteriosus	Persistent ductus arteriosus	Persistent ductus arteriosus, primum atrial septal defect	Persistent ductus arteriosus, bicuspid aortic valve, mitral stenosis

observations. Hernandez, Miller, and Schiebler (1969) reported the rarity of coarctation of the aorta in the American Negro. They offered two plausible reasons for the low incidence. We are inclined to accept their second theory – that the low incidence is a manifestation of poor detection. Sakakibara (1964) also confirmed that the low incidence of congenital heart disease (and coarctation of the aorta in particular) in Japanese infants was related to a delay in diagnosis and referral by practitioners; many patients reached the hospital too late for treatment.

We believe that the apparently low incidence of coarctation of the aorta in Asia and Africa is due to lack of awareness of the lesion and that in many patients the diagnosis is not made as they are symptom free until late adult life. This is in keeping with the findings in our first group of patients, which suggests that coarctation of the aorta occurs much more frequently in White subjects and gives the impression that there is a lower incidence in the African. This impression is a result of case selection. The White, Coloured, and many of the Indian subjects in Natal live in urban areas where they are exposed to a higher standard of informed medical care and where the diagnosis is made more frequently.

A second important clinical observation relates to the fact that coarctation of the aorta in association with other congenital cardiac malformations is frequently responsible for heart failure and death in infancy. The associated malformations often complicate the clinical diagnosis and the lesion is undetected unless special investigations are performed. To determine the true incidence of coarctation of the aorta in a population group, a series of patients should include a large proportion of younger subjects and infants. In clinical reports from various centres in Asia (where this information is given), the percentage of infants included in each study is small. It is likely that many patients with coarctation of the aorta had already died and were therefore excluded. This is in contradistinction to a comprehensive necropsy study from Singapore (Muir, 1960), which gives an incidence of 4·6 per cent for coarctation of the aorta; this is a much higher figure than in the comparable clinical series. Sakakibara (1964) also confirms the high mortality in Japanese infants. We believe that the relative frequency of coarctation of the aorta has been underestimated in Asia and Africa, and that the malformation is often undetected both in clinical and necropsy studies.

Our statistics are not comparable in the different racial groups so that it is not possible to describe the absolute incidence of coarctation of the aorta in each group. Our observations support the hypothesis that the clinical diagnosis is often not made in the Bantu, particularly in young infants, but the relative number of older subjects and the high incidence in the necropsy series suggest that the lesion is not diagnosed. The majority of Bantu subjects live in rural areas where the pressure on medical care is great, and many attending physicians are unaware of the true incidence of the lesion and therefore do not routinely examine the femoral pulses.

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8	*9*	*10*	*11*	*12*
1 mth Persistent ductus arteriosus ventricular septal defect, single atrium	2 mth Persistent ductus arteriosus, aortic stenosis, atrial septal defect	Infancy Persistent ductus arteriosus, ventricular septal defect	8 mth Atrial septal defect small	5 yr Ventricular septal defect

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Requests for reprints to Professor Mervyn Gotsman, Cardiac Unit, Wentworth Hospital, P.B. Jacobs, Durban, Natal, South Africa.